Washington State Department of Health

Fact Sheet

Creutzfeld-Jakob Disease (CJD)
May 2008

What is Creutzfeld-Jakob disease?

Creutzfeldt-Jakob disease (CJD) is the most common human prion disease. Prion diseases are a group of rare brain and nervous system diseases that affect humans and some kinds of animals. They are also known as transmissible spongiform encephalopathies (TSEs). Prion diseases are not caused by germs (i.e., viruses, bacteria). Instead, they are a result of normal brain proteins changing shape and folding into abnormal forms. In affected people, clumps of prion proteins cause brain damage and death.

CJD occurs worldwide in about one to two persons per million each year. In Washington State it affects about six people annually. In all cases, the disease is fatal.

Who is at risk for being affected by CJD?

In the United States, CJD most often occurs in people between 55 and 75 years old. CJD is not spread from person to person. In some cases, 5-15 percent, there is a genetic cause for CJD, but in most cases (about 85 percent), no cause can be determined. When no cause can be determined, the disease is called "sporadic" CJD. Very rarely, CJD has been spread during neurosurgical procedures or human-derived hormone therapies (no longer in use).

Another form of CJD, called variant CJD or vCJD, has affected more than 200 people. Most of those affected were 15-55 years old and lived in or visited the United Kingdom at some time during an epidemic of "mad cow disease" that occurred from1980-1996. Mad cow disease is a cattle prion disease also known as bovine spongioform encephalopathy (BSE). The cause of vCJD is probably consumption of meat products from BSE-affected cattle. There have not been any cases of vCJD acquired in the United States. There are many measures in place to prevent BSE in cattle and to protect the food supply from contamination.

What are the symptoms of CJD?

People with CJD develop dementia and quickly deteriorate mentally. Involuntary twitching, rigid or spasming muscles, lack of coordination and balance, and visual problems are also common symptoms. CJD always leads to death. Approximately half of affected people die within six months of the time symptoms begin, and 90 percent succumb within twelve months.

How is CJD diagnosed?

Symptoms, family history, a lab test, and brain tests such as EEGs and MRIs can help healthcare providers determine that a patient may have CJD. However, the definite diagnosis of CJD requires testing of brain tissue most commonly collected after death in an autopsy. When a person has an illness that could be CJD, it is important that the person's family talk to their physician about getting a clear diagnosis after their loved one's death. There is no charge for the autopsy, transport of the body back to the family, or specialized laboratory testing. Tests are performed at the National Prion Disease Pathology Surveillance Center www.cjdsurveillance.com

How is CJD treated?

There is currently no treatment for CJD. The goal of care is to make the patient comfortable before death.

To support patients' families, the CJD Foundation operates a national toll-free line at (800) 659-1991 and a Web site: http://www.cjdfoundation.org/

For more information call your local health department or Communicable Disease Epidemiology (206) 418-5500 or toll-free 877-539-4344.